What's the story behind the cysts?

A case presentation by

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History

- 47 yr. old black male with a history of back pain for the five months.
- He attributed this pain to heavy lifting and has had good pain relief from aspirin therapy but not from percocets.
- He then developed left-sided scrotal pain and swelling. A testicular ultrasound revealed a left reactive hydrocele.

History

- He was placed on Septra and his provider obtained a chemistry that revealed a creatinine of 1.6mg/dl.
- A follow-up creatinine remained elevated 2 weeks later, and a renal ultrasound was obtained.

Past Medical History/ROS

- Hypertension, untreated, noted to have worsened over the last year.
- Hyperlipidemia, arthalgias (bilateral knees)
- History of dyspnea on exertion
- Decreased visual acuity in right eye (secondary to arteriosclerosis per the patient)

Social History

- Works as sandblaster on propane tanks, with lots of heavy lifting and he takes respiratory precautions
- Smokes 1-2 pks/day
- Patient quit drinking 1 year ago, had long history of alcohol abuse prior to this.

Family History

- Father and paternal aunt died with unspecified kidney problems
- Patient has 11 siblings, two brothers are currently on dialysis.
- One brother has had nephrectomy for unclear reasons
- Another brother is on dialysis and has a history of cystic renal disease

Initial Laboratory Data

- Urinalysis
- Notable for 3-4 nondysmorphic RBCs, 3-4 WBCs, and 0-1 granular casts per /hpf
- Initial Chemistry Na 143 K 4.2 Cl 105 C02 24 BUN 19 Creatinine 1.6
- Initial CBC 6.8> 9.2/27.9 < 227 MCV of 87
- Iron panel showed Fe 29 ug/dl, TIBC of 199 ug/l and Ferritin of 1461

Renal Ultrasound

- Marked dilatation of left renal collection system, bilateral hydronephrosis, and multiple symmetric perph. cortical cysts.
- Renal size was 14cm bilaterally

LNG RT KID
4V2
H4.0MHz
120mm
Abd-Average
Pwr= 0dB MI=1.1
65dB T1/ 0/3/2
Gain= -4dB Δ=2
Store in progress

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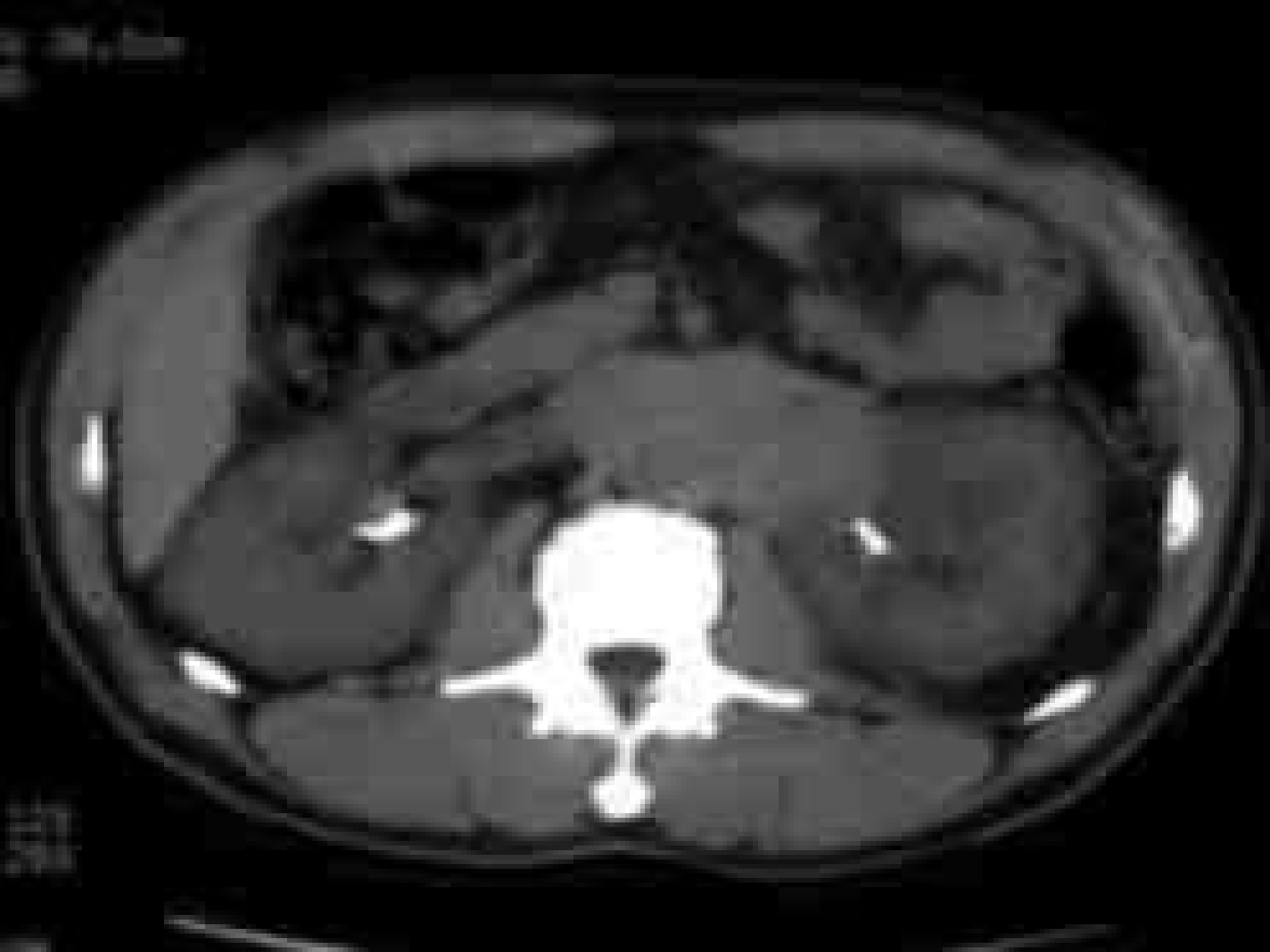
Renal-Lasix Scan

- Angiographic phase demonstrated severe delayed and diminished flow to left kidney, with normal flow and perfusion to right.
- Parenchymal phase was markedly abnormal on left with normal appearance of right (spilt function was 7% on left, and 93.1% on right)
- Tmax on left was 23.5 minutes, right 10 min.

Follow-Up CT Scan

- Bilateral hydronephrosis left> right.
- Multiple cystic lesions bilaterally
- Hypoattunated lesion and perirenal mass with possible renal vein invasion.



Hospital Course

- Pt creatinine continued to rise daily, until it peaked at 3.5 mg/dl (4 days after the initial work-up.
- He continued to have 1.5 liters of urine output a day
- He had low grade fevers throughout the day

Hospital Course

- He underwent CT guided biopsy of the mass , which was unfruitful
- Patient had cystoscopy with bilateral stent placement and creatinine returned to baseline after 5 days to 1.5 mg/dl.
- Intraoperative biopsy of retroperitoneum, noted to have bilateral prominent cystic lesions

BIOPSY

- Dense fibroconnective tissue with no evidence of an epithelial malignancy.
- Evidence of acute and chronic inflammation and prominent blood vessels.
- Simple cysts
- c/w retroperitoneal fibrosis

IDIOPATHIC RETROPERITONEAL FIBROSIS

- First described in 1905 by French urologists
- 1948 Ormond published his accounts of 2 cases , thereafter acknowledged as Ormond’s disease
- Since the late 1970s commonly referred to as idiopathic retroperitoneal fibrosis

CLINICAL FEATURES

- Incidence is 1 in 200,000
- Male predominance of 2-3 to 1
- Usually presents in fifth or sixth decade
- Unusual before 20yrs or after 70 yrs

SYMPTOMS

- Vague, non specific
- Dull, non-localized back or flank pain
- Pain relieved by aspirin and not narcotics
- Weight loss, anorexia, nausea
- Hypertension, and fevers (48%)

Symptoms of advanced disease

- Oliguria/anuria (1/3 have non-functioning kidney at presentation)
- Lower extremity edema
- Scrotal swelling/hydrocele formation
- Bowel obstruction
- Jaundice

Laboratory abnormalities

- Azotemia
- Anemia (normocytic,normochromic)
- Elevated ESR
- Leukocytosis
- Elevated alkaline phosphatase
- Normal urinalysis or microscopic Hematuria

Gross Pathology

- Dense , white, fibrous plaque
- Arises at aortic bifurcation, spreads in perivascular distribution
- Extends laterally to ureters and psoas muscles
- Localized to retroperitoneum
- Can extend outside of retroperitoneum

Histology

- Early stages: Inflammatory cells interspersed amongst fibroblasts and collagen bundles.
- Highly vascular
- Late Stage: relative avascular and acellular, calcifications amidst collagen bundles
- Malignant fibrosis ; scattered nests of well-differentiated malignant cells within the inflammatory infiltrate.

Histology

- Ureters have edema and lymphocyte infiltration
- Underlying aorta has severe atherosclerosis and an associated chronic inflammatory infiltrate
- Lymphatics can be completely obliterated within the mass
- Vessels often have fibrous intima

ETIOLOGY

- Majority of cases are idiopathic (67.8%)
- Clear association with methysergide (13%)
- Associations with beta blockers
- Associations with abd. aortic aneurysm
- Malignancy
- ?Association with systemic collagen-vascular diseases

Possible Associations

- Asbestos
- HLA-B27
- Abdominal injury
- Anticardiolipin antibodies
- Hydralazine, methyldopa

Systemic Fibrosing Disease

- Fibrous psuedotumor of orbit
- Riedel's thyroiditis
- Sclerosing cholangitis
- Mediastinal Fibrosis
- (8-15% have additional fibrosis outside of retroperitoneum)

Periaortic Fibrosis

- Initial fibrosis usually located in area of aorta with most severe atherosclerosis
- Ceroid, polymer of oxidized lipids and proteins
- Recognized by lymphocytes and antibodies once aortic media is invaded
- Association with abdominal aneurysms

Parums, et al. Arch Pathol Medicine 1990

- 20 patients with chronic periaortitis compared to age-match controls
- ELISHA revealed 17/20 with antibody to ceroid vs. 10/20 in age match controls
- Not present in young healthy controls

Methysergide-related Fibrosis

- First described by Graham in 1964
- Case reports of association with other ergot derivatives (bromocriptine)
- Methysergide is also associated with fibrosis elsewhere
- Acts as serotonin antagonist
- Also acts as vasoconstrictor

Malignant Fibrosis

- Desmoplastic response of metastatic tumor
- Displaces the ureters laterally
- Associated with Hodgkin's, lymphomas,sarcomas
- Associated with numerous carcinomas(breast, lung, stomach, colon, kidney, bladder, prostrate)

IMAGING/IVP

- Classic triad includes unilateral (20%), bilateral (68%) hydronephrosis
- Medial deviation of middle one third of ureters (versus lateral deviation in most neoplasms)
- tapered narrowing of the ureters at L4/5
- Retrograde pyelography : nondistendable ureters

IMAGING CONTINUED

- Lymphangiography; delayed passage of contrast through iliac and paraaortic lymphatics,
- Obstruction at L3-4
- small defects within the mesenteric and paraaortic lymph nodes

CT Scan

- Modality of choice
- Aorta is not displaced anteriorly
- Ureters displaced medially
- No local bone destruction
- Can not distinguish idiopathic from malignant retroperitoneal process

Other modalities

- Ultrasound (homogenous appearance)
- MRI: better vascular definition
- T1 and T2 images are distinct from adjacent tissue
- Can differentiate early versus late stages of fibrosis
- Cannot distinguish between benign and malignant process

Diagnosis/Biopsy

- CT scan-guided fine needle aspiration less effective, secondary to small tissue cores
- Several reports of delayed diagnosis of malignancy
- Multiple deep retroperitoneal biopsies is gold-standard
- No randomized studies for comparison

Treatment

- Primary treatment is surgical
- Goal is to establish diagnosis
- Exclude malignancy
- Restore renal function
- Discontinue offending agents
- Review medical therapies available

Ureterolysis

- Multiple deep biopsies of retroperitoneum
- Ureterolysis
- Spontaneous contralateral improvement
- Transposition of ureters to intraperitoneal position, or laterally
- Wrapped with omental fat, Gor-Tex graft
- Renal autotransplantation

Tipgraft, et al. Br J Urology 1982

- Follow-up of 21 patients treated with ureterolysis and either omental wrapping of ureters or intraperitoneal placement
- Recurrence noted in 2/21 patient with omental wrapping
- Recurrence noted in 2/3 patients with intraperitoneal placement

Steroids

- Strong evidence of immune-mediate phenomenon
- Acute resolution of edema
- Bridge to surgery
- Adjuvant therapy to surgery
- Used if recurrence post operatively

Higgins,et al
Br j Surgery, 1988

- Collection of case studies of 17 patients
- Steroids given initially 80mg po qd, tapered to 20mg po qd
- Noted restoration of renal function in 15/17
- High incidence of recurrence once steroids stopped
- No long term follow-up

Other immunomodulating agents

- One case report of treatment with methotrexate therapy
- Azathioprine, doses of 100-150mg mg po qd (used alone or after pulse steroids)
- One case report of cyclophosphamide use

Harreby,et al Scan J Urol Nephrol, 1994

- 11 patients, biopsy proven retroperitoneal fibrosis
- Pulse methylprednisolone (3gms over 3 days)
- Followed by azathioprine or penacillamine 450mg po qd
- 8/11 had restoration of renal function

Tamoxifen

- Case reports of use
- Retroperitoneal fibrosis has similar histologic features as desmoid tumors
- Prompt recurrence after stopping the medication
- Mechanism is unclear

Conclusions

- Have a high index of suspicion
- Ultrasound can be insensitive
- Primary treatment is surgical
- Can be part of a systemic fibrosing disorder with a significant inflammatory component, perhaps amenable to immunosuppressant regimens
- We still can’t explain the cysts (sorry!)